

CASE REPORT

Howell-Jolly bodies on peripheral smear leading to the diagnosis of congenital hyposplenism in a patient with septic shock

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Key Clinical Message

We present a case of isolated congenital hyposplenism that was discovered after the peripheral smear revealed Howell-Jolly bodies. This case serves as the basis for a review of hyposplenism for the general practitioner.

Keywords

Howell Jolly Bodies, hyposplenism, immunocompromised, peripheral smear.

Introduction

The peripheral smear is a powerful tool in the arsenal of every hematologist, as well as the general internist. Here, we present a case that relied on the peripheral smear to arrive at a rare diagnosis of congenital hyposplenism. While acquired hyposplenism, as well as congenital asplenia, have been well documented, congenital hyposplenism is not described in the literature. Therefore, our case is notable for several reasons. First, it highlights the medical anomaly of congenital hyposplenism. Second, it highlights the importance of the peripheral smear in making a diagnosis of hyposplenism. Finally, these findings led to an intervention that may prevent future death or morbidity in this patient.

Case Presentation

A 45-year-old woman was transferred to our urban, tertiary care hospital from a local community hospital for management of septic shock secondary to pneumococcal meningitis. The patient initially described vague, mild

headaches, and myalgias that ultimately progressed into severe bitemporal headaches, fever, and neck pain. Significant medical history included cervical cancer treated with hysterectomy 14 years prior and chronic sinus infections. On transfer to our hospital, her complete blood count revealed a white blood cell (WBC) count of 25.7 K/UL, hemoglobin of 10.6 g/dL, and a platelet count of 149 K/UL. The patient also had multiple metabolic derangements including hypocalcemia (6.1 mg/dL), hyperglycemia (585 mg/dL), as well as a mild coagulopathy (INR = 1.78 and PTT = 41 s). HIV testing was negative. Lumbar puncture demonstrated purulent fluid consistent with bacterial meningitis, though CSF gram stain and culture were negative. A peripheral smear was reviewed and it demonstrated significant Howell-Jolly bodies (Fig. 1). On workup for the etiology of these findings, CT of the chest, abdomen, and pelvis with contrast was notable for markedly reduced spleen size, but without irregularities suggestive of intrinsic splenic pathology (Fig. 2). The patient was successfully treated with hemodynamic support and intravenous antibiotics. After recovering from the critical phase of her illness, in

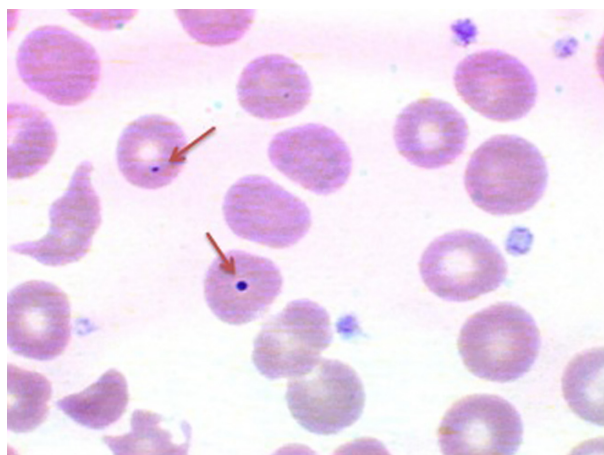


Figure 1. 400× view of peripheral blood smear demonstrating Howell-Jolly bodies.

light of her hyposplenism, she received vaccinations for pneumococcus, meningococcus, and hemophilus influenzae. A repeat blood count after resolution of her infection showed that her platelets had risen to 913 K/UL, consistent with her hyposplenism.

Discussion

While hyposplenism itself is a relatively common occurrence, healthcare providers are less familiar with the possibility of congenitally reduced splenic size, without previous infarct/insult. Familiarity with the presentation, peripheral smear findings, and management of congenital hyposplenism is critical to appropriate patient care.

Splenic function and the peripheral smear

The spleen has two main functions: the filtration of deranged red blood cells, and defense against infections. Thus, with inadequate splenic function, patients are predisposed to increased frequency and severity of infections, specifically with encapsulated bacterial organisms.

Peripheral smear findings in patients with a dysfunctional spleen include the presence of Howell-Jolly bodies, thrombocytosis, lymphocytosis, and monocytosis [1]. Howell-Jolly bodies are small, intra-erythrocytic remnants of erythrocyte nuclei. These inclusions are solitary in each erythrocyte and strongly basophilic. These are often confused with overlying platelets, but can be distinguished by the presence of a “halo” around overlying platelets. A normally functioning spleen removes the remnants from erythrocytes, so their presence is pathognomonic for splenic dysfunction.

In the presented case, it was the finding of Howell-Jolly bodies on the peripheral smear that led to the suspicion

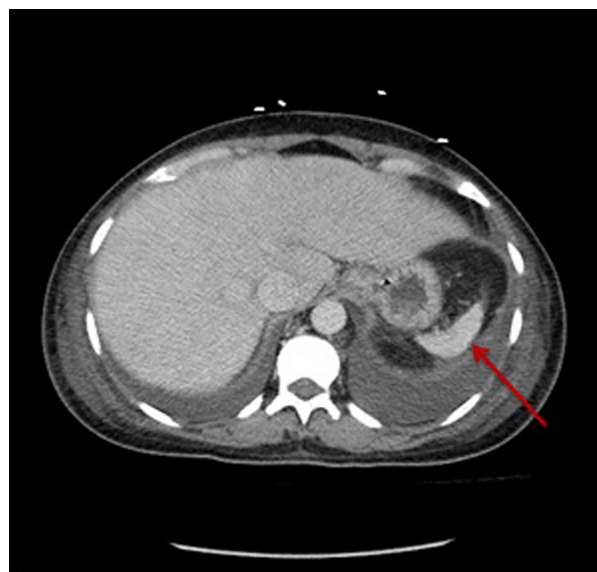


Figure 2. Transverse image of markedly reduced spleen size (arrow).

of poor splenic function as the underlying etiology for her septic shock.

Radiographic findings in congenital hyposplenism

Most commonly, splenic abnormalities present as complete absence of the spleen. Alternatively, a small, pathologic appearing spleen may be visualized on imaging. With the latter, the spleen typically demonstrates evidence of infarction or infiltration. On abdominal CT, our patient demonstrated a remarkably small spleen, but no evidence of pathology (Figure 2).

Etiology and clinical presentation of asplenia and hyposplenism

The clinical presentation of a patient with asplenia or hyposplenism is related to both the etiology of the splenic dysfunction, as well as infectious complications. Asplenia is most frequently found after splenectomy for hematological, immunological, or traumatic indications [2]. Rarely, asplenia can be a congenital condition [3, 4]. Hyposplenism can be found in many hepatic, gastrointestinal, hematologic, autoimmune, and infectious disorders [1]. Most notably, all patients with sickle cell anemia (SCA) will accrue multiple splenic infarcts, which can result in reduced splenic size and hyposplenism [5]. Specific disease leading to hyposplenism are Celiac disease, HIV infection, alcoholic liver disease, as well as being found in patients undergoing bone marrow transplantation.

Rare congenital conditions may result in reduced spleen size and decreased function. Disease such as Autoimmune Polyendocrinopathy Candidiasis Ectodermal Dystrophy (APECED), Ivemark Syndrome and Stormorken Syndrome present with various constellations of pathologic organ development including splenic agenesis or dysgenesis. Rarer still is isolated congenital asplenia [6, 7]. Cases describing congenital *hyposplenism*, as seen in our case, are not represented in the peer-reviewed literature.

Inadequate splenic function can lead to a relatively immunocompromised state, which may lead to infections referred to as “overwhelming post-splenectomy infections” (OPSI). OPSI will often present with a flu-like prodrome and may progress in severity to septic shock. The most common culprits are encapsulated organisms specifically *Streptococcus pneumonia*, *Neisseria meningitides*, and *Hemophilus influenza type B*.

OPSI are most commonly seen in individuals under the age of 2 years, as these patients are typically not yet immunized. However, presentations can occur in adulthood [8]. This was the case in our patient described above, who was middle age at time of presentation. While she had no history of overwhelming sepsis, on further review, she did report frequent illnesses in childhood[†], specifically bacterial sinusitis infections. Additionally, her ultimate presentation was preceded by a prodrome consistent with OPSI presentation.

Management of a patient with congenital hyposplenism

In clinical practice, patients with hyposplenism should be approached in the same manner as asplenic patients. Specifically, patients with hyposplenism should receive vaccinations against encapsulated organisms, including streptococcus, meningococcus, and hemophilus influenzae, as well as diligent administration of zoster, MMR and other age appropriate immunizations [9, 10]. Furthermore, asplenic patients should be given antibiotics to take at the onset of fever over 100.4 degrees Fahrenheit (38.4 Celsius), and should then proceed immediately to the nearest emergency department. Common antibiotics are amoxicillin-clavulanate, cefuroxime, or moxifloxacin (in patients with serious penicillin allergy) [6]. Prophylactic daily antibiotics are routinely given to asplenic and hyposplenic children under the age of five, but evidence is lacking in adults. Perhaps the most critical intervention may be to ensure that patients receive appropriate education on the risks of hyposplenism and the importance of

seeking immediate medical care in the event of a fever [9, 11, 12].

Our patient did not have a known history of hyposplenism, and, thus, was not appropriately vaccinated against encapsulated organisms prior to presentation. Once she recovered, though, she received vaccination against streptococcus, meningococcus, and hemophilus influenzae. Additionally, she was provided with antibiotics to take at the onset of fever. Her daughter was evaluated in our outpatient hematology clinic for education regarding the possibility of congenital hyposplenism.

Conclusion

This case highlights an important peripheral smear finding that led to the discovery of hyposplenism as the underlying cause of our patient's overwhelming sepsis. Once the patient recovered, the diagnosis of hyposplenism led to several interventions that will decrease the risk of future episodes of severe sepsis.

While acquired hyposplenism and congenital asplenia have been well documented in the literature, congenital hyposplenism is less described. This case highlights an important finding on the peripheral smear that can lead to interventions that may prevent future episodes of severe sepsis in hyposplenic patients.

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[†]Correction added on 16 July 2015, after first online publication: ‘childhood’ and ‘illnesses’ were transposed.

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